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Multiple secondary neoplasms in nevus sebaceus excision

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ABSTRACT

Nevus sebaceus is a congenital hamartoma associated with several secondary tumors. We report the case of a 19-year-old woman who presented with changes in a nevus sebaceus lesion on her parietal scalp, which was subsequently excised. Upon pathological examination, both basaloid hamartoma and syringocystadenoma papilliferum were noted within the specimen, which is rare. The primary treatment modalities for nevus sebaceus are either close clinical observation or surgical excision, but no definitive consensus exists on the excision timeline of nevus sebaceus.

KEYWORDS Basaloid hamartoma; dermatopathology; hormonal changes; nevus sebaceus; scalp lesion; syringocystadenoma papilliferum; tumor

Nevus sebaceus, also referred to as nevus sebaceus of Jadassohn and organoid nevus, is a congenital hamartoma composed of follicular, sebaceous, and apocrine structures and is associated with a number of secondary tumors of varying rarity. These lesions are primarily found on the head or neck, most notably the scalp.[1] Approximately 0.3% of newborns are affected by nevus sebaceus, and the lesion does not show preference for gender or race.[2] The lesion typically grows slowly with age, with the most rapid growth occurring during the pubertal hormonal surge.[1] The two most common secondary neoplasms developing within a nevus sebaceus are trichoblastoma and syringocystadenoma papilliferum during or after puberty.[3] Though other secondary neoplasms can and do occur within nevus sebaceus, it is extremely unusual to observe two distinct secondary tumors at once.

CASE REPORT

A 19-year-old woman presented to the clinic with a changing lesion on the right parietal scalp. She stated that the lesion has been present since birth but over the last several years had started to change. The risks of secondary tumor development within the lesion were discussed with the patient as well as potential surgical removal. She elected to have the lesion excised via elliptical excision with margins. *Figure 1a and Figure 1b* display the patient's scalp before and after the lesion was removed. Note the linear blaschkoid hairless orange-yellow plaque on the scalp. The history of becoming more verrucous around puberty is a classic presentation. In this particular case, both basaloid hamartoma and syringocystadenoma papilliferum were noted within the excision specimen *(Figure 1c, 1d)*.

DISCUSSION

Nevus sebaceus is of particular clinical interest because of its association with the development of secondary neoplasms of varying rarity. Secondary tumors are typically benign, but malignant neoplasms have also been observed. The most common benign neoplasms include trichoblastoma and syringocystadenoma papilliferum. Additional benign neoplasms such as apocrine/eccrine adenoma, trichilemmoma, and sebaceoma have been observed.[3] It has been estimated that approximately 40% of syringocystadenoma papilliferum lesions are associated with nevus sebaceus.[4]

The risk of a secondary carcinoma developing within a nevus sebaceus is low. Apocrine carcinoma, squamous cell carcinoma, and infundibular cyst have all been observed as secondary tumors found within nevus sebaceus but at a much lower prevalence. Normally the tumors associated with nevus sebaceus are benign, with the most common malignant tumors being squamous cell carcinoma, basal cell carcinoma, and sebaceous carcinoma.[5]

Though multiple neoplasms can be observed within a nevus sebaceus lesion, it is quite rare to observe both basaloid hamartoma and syringocystadenoma papilliferum together. However,

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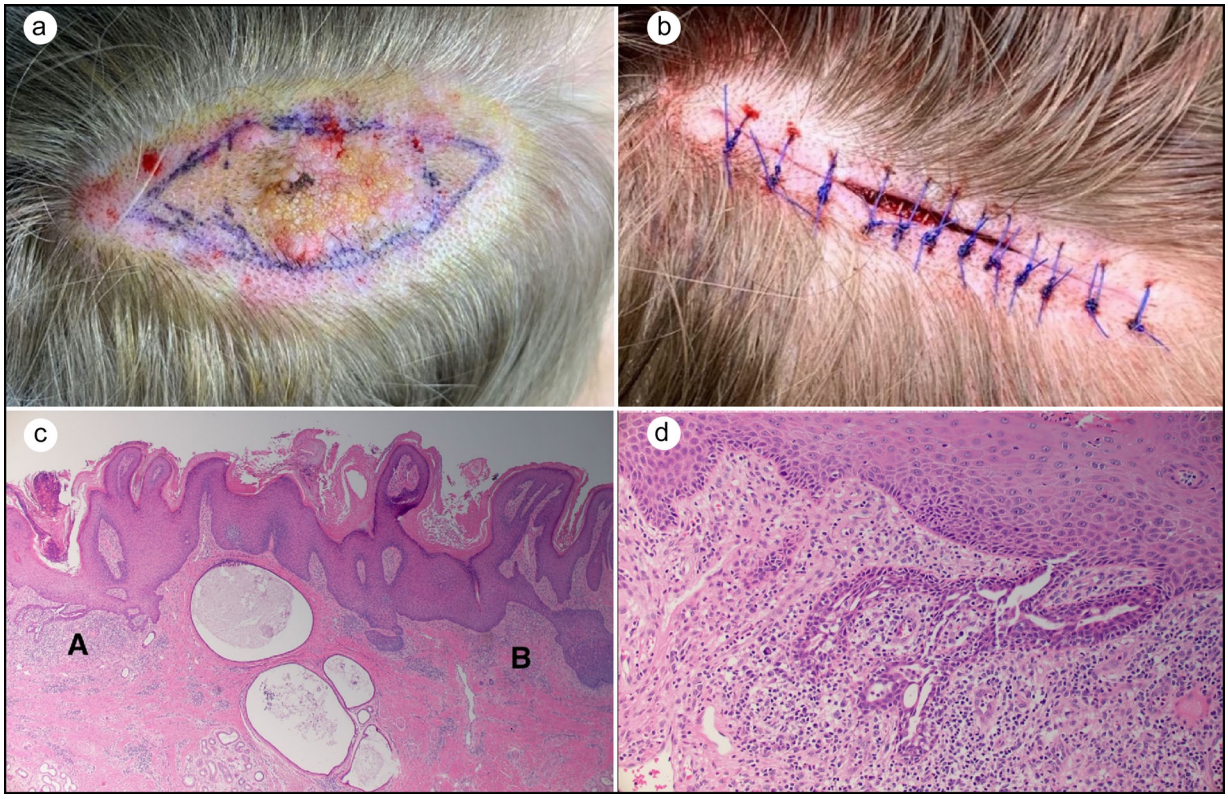

Figure 1. Nevus sebaceus lesion on the patient's scalp **(a)** prior to excision and **(b)** immediately following excision. **(c)** Epidermis showing irregular acanthosis, hyperkeratosis, and papillomatosis. A small syringocystadenoma (A) and basaloid hamartoma (B) are present along with prominent apocrine glands. **(d)** Syringocystadenoma papilliferum consisting of ductal structures extending from the epidermis with surrounding plasma cell inflammation.

our review of the literature revealed a single case report describing six unique neoplasms found in a single lesion.[6] Malignant secondary tumor transformation has been observed almost exclusively in adults, with a recent retrospective analysis of 707 cases showing that patients over the age of 18 account for 96% of transformations, with the remaining 4% of cases among patients aged 11 to 17.[3]

The primary treatment modalities for nevus sebaceus are either close clinical observation or surgical excision, but no definitive consensus exists on the excision timeline of nevus sebaceus. Some suggest that the complications of excision are more prevalent during childhood and thus surgical excision should be delayed until adulthood if possible.[7] Others suggest that excisions should be performed during infancy, while the tumors are relatively small and scar management is most favorable.[8] Though it is true that small childhood lesions are more favorable for excision, other factors such as tolerability of local and general anesthesia must be accounted for.[1] Prophylactic surgical excision of nevus sebaceus has been recommended in the past due to the conflicting proportions of malignant tumors reported in the literature and potential misclassification of trichoblastoma as basal cell carcinoma, thus increasing the reported incidence of malignant neoplasms.[9] Accounting for all conflicting rationales has led to the majority of surgeons adopting the approach of excision prior to pubertal enlargement when local and general anesthesia can be well tolerated vs observing until malignant features develop.

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